Membranous atresia of anomalous left coronary artery originating from the pulmonary artery presenting as an intravascular tumor

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We present the case of a 31-year-old woman with anomalous left coronary artery arising from the pulmonary artery (ALCAPA) presenting as a mass within the pulmonary artery. The mass was found to be a complex cystic structure circumferentially attached to the left coronary ostium, probably as a result of membranous atresia of the left coronary ostium. Informed consent and approval from the Froedtert Hospital institutional review board were obtained for this report.

CLINICAL SUMMARY

In the course of an emergency department workup of a 31-year-old woman, 130 kg in weight, with chest and flank pain, 12-lead electrocardiographic and cardiac enzyme results were negative. Computed tomography (pulmonary embolism protocol) of the chest revealed a 3-cm complex enhancing mass of the pulmonary artery near the valve (Figure 1). Pain resolved after treatment for a urinary tract infection; however, she continued to report dyspnea on exertion. Cardiac magnetic resonance imaging yielded concern for myxoid sarcoma or angiosarcoma and demonstrated normal left ventricular systolic function without perfusion abnormality. Coronary branching was not well delineated. Systematic workup for metastatic disease yielded negative results.

In the operating room, results of transesophageal echocardiography were consistent with a complex cystic mass. Inspection of the heart demonstrated a mildly enlarged right coronary artery. Near the completion of standard antegrade cardioplegia administration, the pulmonary artery was opened distal to the mass, revealing a 2- to 3-cm intact cystic structure containing dark fluid. Frozen sections led to concern regarding a myxoid tumor. In proceeding with pulmonary root resection, abnormally thickened tissue was encountered in a fat pad along the left side of the proximal pulmonary artery. Exploration of the interior of the mass demonstrated a single ostium and vessel originating at the commissure of the posterior left anterior sinus. Exploration of the aortic root demonstrated only a right coronary ostium. Because of the concern for malignancy and need for wide margins, the pulmonary root was harvested leaving only 1 mm of left main coronary artery (specimen shown in Figure 2). A generous doughnut-style hood of porcine extracellular matrix patch was anastomosed to the left main coronary artery, with the outer circumference

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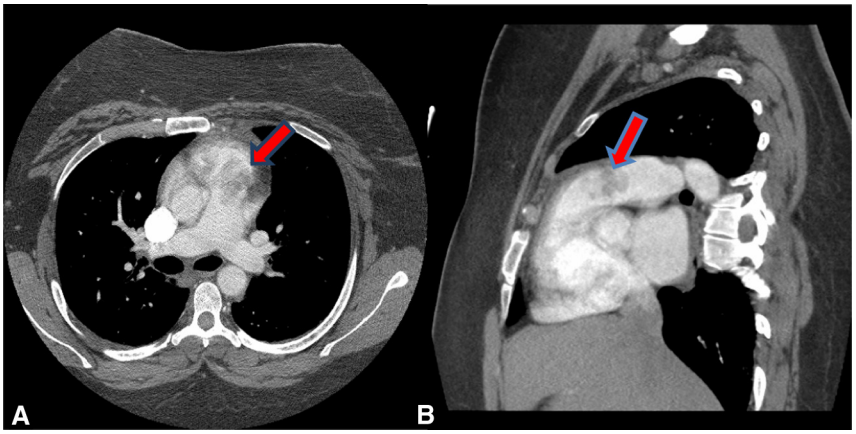

FIGURE 1. Computed tomography demonstrating an intravascular mass of the pulmonary artery near the valve (*red arrows*) as seen on transverse (A) and sagittal (B) views.

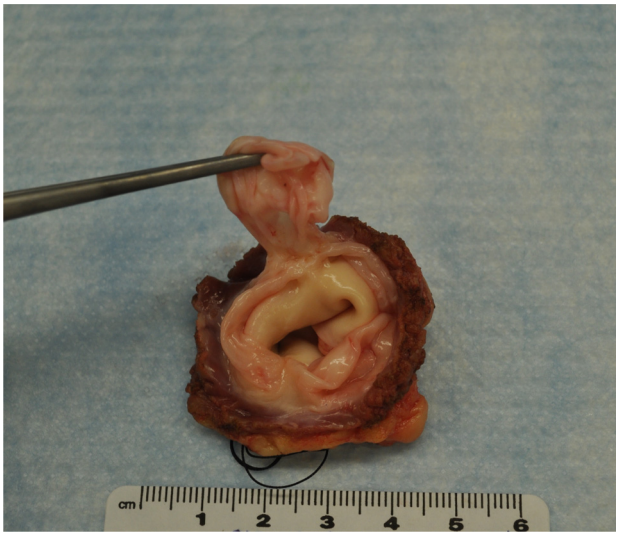

FIGURE 2. Excised pulmonary root as viewed from the proximal aspect with the cystic mass decompressed (being grasped by forceps).

anastomosed to a 6-mm Dacron polyester fabric graft interposed to the left coronary sinus of the aorta. The pulmonary root was replaced with a 30 mm ABO-compatible pulmonary homograft. The patient had an uneventful recovery and was discharged home on postoperative day 6.

Cardiac-gated computed tomography and echocardiography 2 months after surgery showed a widely patent interposition graft with antegrade flow, normal function, and a competent pulmonary valve. Final pathologic examination of the cystic mass demonstrated no malignancy and cells with surface markers consistent with endothelial lineage. Clinically, the patient continues to do well 7 months after hospital discharge.

DISCUSSION

We present an extremely rare case of what we suspect represents membranous atresia of the ostium of an ALCAPA preventing coronary decompression and ischemic findings. Long-term survivors of ALCAPA are thought to present ultimately with signs of myocardial ischemia, evidenced by left ventricular subendocardial fibrosis on echocardiography, and 90% of these patients experience sudden cardiac death at a mean age of 35 years.[1-3] We suspect that in our patient the intact complex cystic structure, which likely resulted from chronic expansion of an original membrane of the left coronary artery ostium, prevented left coronary artery decompression and the development of frank ischemic changes. To our knowledge, this has not previously been reported in the literature. Observations supporting (but not proving) this interpretation include (1) markers of endothelial lineage on final pathologic report and no evidence of malignancy; (2) no preoperative evidence of ischemia; (3) good aortic antegrade arrest, which did not require exceptionally frequent readministration of cardioplegia; (4) no leaking from a tense cystic structure with antegrade cardioplegia; and (5) only mildly enlarged right coronary artery without an extensive network of large collaterals around the base of the aorta and pulmonary artery. In retrospect, better appreciation of the import of mild focal delayed enhancement of the cardiac apex on cardiac magnetic resonance imaging could have prompted a coronary-specific magnetic resonance imaging or computed tomographic protocol and potentially a correct preoperative diagnosis.

The decision to proceed according to oncologic concerns was perhaps overly influenced by interpretations of preoperative studies, results of frozen section studies, and the consequence of proceeding with isolated therapy for ALCAPA in the event of ultimate confirmation of malignancy. We chose a modified Cabrol technique to establish an antegrade 2-coronary system. We acknowledge concerns related to circumferential patch material and plan to monitor the patient accordingly. Bypass grafting, an acknowledged alternative, remains a viable option for this patient should graft stenosis occur. In general, we do not advocate ligation and a single-coronary repair because of potentially inferior long-term results.[4,5]

References

1. Berdjis F, Takahashi M, Wells WJ, Stiles QR, Lindesmith GG. Anomalous left coronary artery from the pulmonary artery. Significance of intercoronary collaterals. *J Thorac Cardiovasc Surg*. 1994;108:17-20.
2. Alexi-Meskishvili V, Berger F, Weng Y, Lange PE, Hetzer R. Anomalous origin of the left coronary artery from the pulmonary artery in adults. *J Card Surg*. 1995;10:309-15.
3. Yau JM, Singh R, Halpern EJ, Fischman D. Anomalous origin of the left coronary artery from the pulmonary artery in adults: a comprehensive review of 151 adult cases and a new diagnosis in a 53-year-old woman. *Clin Cardiol*. 2011;34:204-10.
4. Dodge-Khatami A, Mavroudis C, Backer CL. Anomalous origin of the left coronary artery from the pulmonary artery: collective review of surgical therapy. *Ann Thorac Surg*. 2002;74:946-55.
5. Wilson CL, Dlabal PW, McGuire SA. Surgical treatment of anomalous left coronary artery from pulmonary artery: follow-up in teenagers and adults. *Am Heart J*. 1979;98:440-6.